which measured 1 cm in diameter (fig 1B). There was less thickening of the other extraocular muscles. A high signal was seen in these muscles on T_2 weighted imaging, indicating oedema and acute inflammation.

A diagnosis of orbital myositis (OM) was made and treatment was started with oral prednisolone 60 mg daily. Her symptoms rapidly improved and steroid treatment was withdrawn within three months. Five months after her diagnosis she had mild restriction of horizontal movements in the right eye and repeat MR scan showed an element of chronic fibrosis.

DISCUSSION

OM is characterised by the onset of painful and limited extraocular movements, diplopia, ptosis, swelling of the lid, and localised chemosis and injection over the insertion of the inflamed muscle.[1]

The most commonly affected muscles are the superior complexes and the medial rectus muscle. OM may attack more than one muscle, and may be bilateral or recurrent.

The major differential diagnosis is thyroid ophthalmopathy. However, dysthyroid myopathy is usually painless in onset, symmetrical, slowly progressive, and associated with systemic manifestations of Grave's disease. Lid retraction, limitation of the movement opposite to the affected muscle, and deterioration of visual function (colour vision, visual field, and visual acuity) may also occur in thyroid eye disease, in contrast with OM. Additional diseases that should be considered in the differential diagnosis include orbital cellulitis, metastasis, Tolosa-Hunt syndrome, trochleitis, and infectious myositis due to trichinosis.[2]

Imaging of the orbit in OM shows diffuse enlargement of the extraocular muscles, which exhibit slightly blurred margins.

Associations with OM include distant inflammatory disease such as Crohn's disease,[3] Lyme disease,[4] and Wegener's granulomatosis.[5] It may also be a manifestation of a paraneoplastic syndrome.[6] Although there are published reports of OM associated with psoriatic arthropathy[7] and systemic lupus erythematosus,[8 9] there has only been one case reported in association with RA.[10] Management is with corticosteroids and the rapid response is almost diagnostic.

Authors' affiliations
S Nabili, Ophthalmology Department, Gartnavel General Hospital, Glasgow, UK
D W McCarey, H A Capell, Centre for Rheumatic Diseases, Glasgow Royal Infirmary, Glasgow, UK
B Browne, Ophthalmology Department, Glasgow Royal Infirmary, Glasgow, UK

Correspondence to: Dr D W McCarey, Centre for Rheumatic Diseases, Glasgow Royal Infirmary, 84 Castle Street, Glasgow G4 0SF, UK; gcl376@clinmed.gla.ac.uk

Accepted 27 May 2002

REFERENCES

1 **Berger JW**, Rubin PA, Jakbiec FA. Pediatric orbital pseudotumor: case report and review of the literature. Int Ophthalmol Clin 1996;36:161–77.
2 **Bouree P**, Bouvier JB, Passeron J, Galanaud P, Dormont J. Outbreak of trichinosis near Paris. BMJ 1979;i:1047–9.
3 **Dunro CA**, Ehrlich R, Taylor R, Buncic JR, Hughes P, Griffiths AM. Keeping an eye on Crohn's disease: orbital myositis as the presenting symptom. Can J Gasteroentrol 1997;11:497–500.
4 **Seidenberg KB**, Leib ML. Orbital myositis with Lyme disease. Am J Ophthalmol 1990;15:13–16.
5 **Pinchoff BS**, Sphalinger DA, Bergstrom TJ, Sandall GS. Extra ocular muscle involvement in Wegner's granulomatosis. J Clin Neurol Ophthalmol 1983;3:163–8.
6 **Harris GJ**, Murphy ML, Schmidt EW, Hanson GA, Dotson RM. Orbital myositis as a paraneoplastic syndrome. Arch Ophthalmol 1994;112:380–6.
7 **Ajitsaria R**, Dale R, Ferguson V, Mayou S, Cavanagh N. Psoriasis, psoriatic arthropathy and relapsing orbital myositis. Clin Exp Dermatol 2001;26:274–5.
8 **Grimson BS**, Simons KB, Orbital inflammation, myositis, and systemic lupus erythematosus. Arch Ophthalmol 1983;101:736–8.
9 **Serop S**, Vianna RN, DeLaey JJ. Orbital myositis secondary to systemic lupus erythematosus. Acta Ophthalmol 1994;72:520–3.
10 **Panfilio CB**, Hernandez-Cossio O, Hernandez-Fustes OJ. Orbital myositis and rheumatoid arthritis: case report. Arq Neuropsiquiatr 2000;58:174–7.

Stiff man syndrome presenting with low back pain

A Bastin, V Gurmin, R Mediwake, J Gibbs, H Beynon

Ann Rheum Dis 2002;**61**:939–940

Stiff man syndrome (SMS) is a rare, disabling neurological disorder characterised by progressive muscle rigidity and painful episodic spasms of the axial and proximal limb muscles. Diagnosis is based on the recognition of typical clinical features and characteristic EMG findings.[1] However, although it is well described, SMS is probably underdiagnosed because of a lack of awareness of its clinical manifestations. It may present to a range of specialties and should be considered in all patients with unexplained back pain, stiffness, and muscle spasms. We present a typical case of stiff man syndrome in a patient who was referred to our unit, having previously been seen by a number of doctors and a neurosurgeon, without a diagnosis.

CASE REPORT

A 42 year old man with a 28 year history of insulin dependent diabetes mellitus (IDDM) and a two year history of progressive pain and stiffness affecting his lower back and abdomen was seen in the rheumatology clinic. He also had painful, intermittent muscle spasms. His functional ability had significantly decreased and he complained of extreme difficulties in performing tasks requiring back flexion. Bladder and bowel function was normal. Blood sugars were controlled with insulin and he had no diabetic complications.

On examination, he had a marked lumbar lordosis, a prominent thoracic kyphosis, and a protuberant abdomen (fig 1). There was pronounced paraspinal and abdominal wall muscle rigidity. Neurological examination was otherwise normal. Blood tests showed a mildly raised haemoglobin A_{1c} (7.4) and plain radiography of the spine confirmed the clinical findings. The EMG was classical for SMS and showed sustained motor unit activity in agonist and antagonist axial and limb muscles, despite the patient's attempts at relaxation. The diagnosis was supported by the detection of antibodies against glutamic acid decarboxylase (GAD) in his serum. His condition only mildly improved with baclofen 80 mg/day, and diazepam has now been introduced.

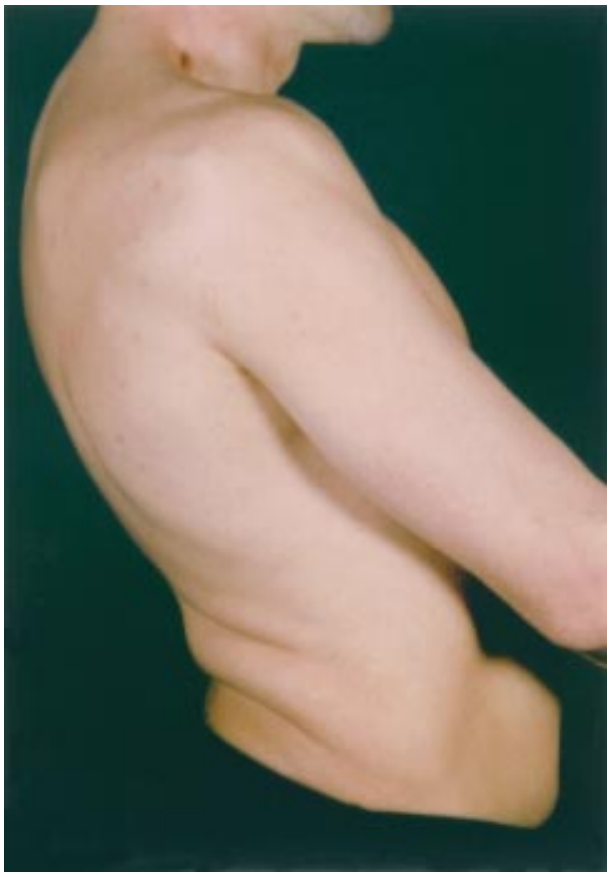

Figure 1 Patient with stiff man syndrome.

DISCUSSION

SMS has an insidious onset, usually in the fourth or fifth decades, with progressive muscle rigidity and episodic spasms affecting the axial and limb muscles. Muscle rigidity can lead to contractures, and simultaneous contraction of the thoracolumbar paraspinal and abdominal wall muscles causes lumbar hyperlordosis. Episodic muscle spasms are classical in SMS and their absence should raise suspicions about the accuracy of the diagnosis.[1] The spasms, which are often provoked by emotional upset or sudden movement, can be extremely painful, generating forces capable of fracturing long bones.[2] Rigidity and spasms gradually impair voluntary movements and postural reflexes, resulting in slow, restricted movements and an increased risk of falls. Intellect is not affected, and motor and sensory nerve examination is also normal. However, almost all patients have an abnormal EMG pattern, which shows continuous motor unit activity in affected muscles.[1]

The cause of SMS is unknown, but an autoimmune pathogenesis is suggested by the presence of autoantibodies and its strong association with autoimmune conditions, such as IDDM and thyroiditis.[3] Antibodies against GAD are present in about 60% of patients with SMS.[4] GAD is the rate limiting enzyme in the synthesis of γ-aminobutyric acid (GABA), one of the main inhibitory central neurotransmitters. Reductions in GABA production may therefore impair transmission at central nervous system inhibitory synapses, resulting in the continuous motor unit activity seen in this disease. Antibodies against amphiphysin, a nerve terminal protein, have been detected in up to 5% of anti-GAD negative patients. They are strongly associated with paraneoplastic SMS and their presence should prompt careful investigation to exclude malignancy, particularly breast cancer.[5] However, approximately 40% of patients have no evidence of autoantibodies, suggesting that the pathogenesis of this syndrome may be heterogeneous.[6]

Drugs that enhance GABA-mediated central inhibition, such as diazepam (up to 300 mg/day), baclofen, sodium valproate, and vigabatrin, are the mainstay of treatment for this previously refractory disease. Immunomodulatory treatments have also been used and favourable responses to corticosteroids, plasmapheresis, and intravenous immunoglobulin have been reported.[7–9] Physiotherapy is also important, offering a valuable adjunct to drug treatment.

SMS should be considered in all patients with unexplained back pain, stiffness and muscle spasms as early recognition and therapeutic intervention can significantly decrease morbidity and improve quality of life.

Authors' affiliations
A Bastin, V Gurmin, R Mediwake, H Beynon, Department of Rheumatology, The Royal Free Hospital, Pond Street, London NW3 2QG, UK
J Gibbs, Department of Neurology, The Royal Free Hospital

Correspondence to: Dr H Beynon; hlcbeynon@aol.com

Accepted 15 March 2002

REFERENCES

1 **Lorish TR**, Thorsteinsson G, Howard FM. Stiff-man syndrome updated. Mayo Clin Proc 1989;64:629–36.
2 **Layzer RB**. Stiff-man syndrome –an autoimmune disease? N Engl J Med 1988;318:1060–2.
3 **McEvoy KM**. Stiff-man syndrome. Mayo Clin Proc 1991;66:300–4.
4 **Solimena M**, Folli F, Aparisi R, *et al*. Autoantibodies to GABAergic neurons and pancreatic beta cells in stiff-man syndrome. N Engl J Med 1990;322:1555–60 .
5 **Folli F**, Solimena M, Cofiell R, *et al*. Autoantibodies to a 128-kd synaptic protein in three women with the stiff-man syndrome and breast cancer. N Engl J Med 1993;328:546–51.
6 **Grimaldi LME**, Martino G, Braghi S, *et al*. Heterogeneity of autoantibodies in stiff-man syndrome. Ann Neurol 1993;34:57–64.
7 **Piccolo G**, Cosi V, Zandrini C, Moglia A. Steroid-responsive and dependent stiff-man syndrome: a clinical and electrophysiological study of two cases. Ital J Neurol Sci 1988;9;559–66.
8 **Brashear R**, Phillips HL II. Autoantibodies to GABAergic neurons and response to plasmapheresis in stiff-man syndrome. Neurology 1991;41:1588–92.
9 **Barker RA**, Marsden CD. Successful treatment of stiff man syndrome with intravenous immunoglobulin. J Neurol Neurosurg Psychiatry 1997;62:426–42.